Bilateral Agenesis of the Internal Carotid Artery Associated with Basilar Artery Aneurysm Treated via the Endovascular Route

A Case Report

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Summary

Bilateral agenesis of the internal carotid artery is a rare anomaly of embryonic development frequently associated with intracranial aneurysm. We describe a case involving an aneurysm that burst in the third middle of the basilar artery and exhibited a bilateral agenesis of the internal carotid artery. The aneurysm was treated via an endovascular route using detachable coils.

Introduction

The absence of the internal carotid artery (ICA) is a rare anomaly of embryonic development (ED) that occurs in less than 0.01% of the population [1,2]. The estimated prevalence of cerebral aneurysm in the general population is between 2% to 4%, but in association with the absence of the ICA, its occurrence ranges from 24% to 34% [1]. We describe a case of subarachnoid hemorrhage (SAH) and saccular aneurysm of the third middle of the basilar artery in a patient with bilateral agenesis of the ICA, which was treated via an endovascular route using detachable coils.

Case Report

A formerly healthy 34-year-old woman presented an intense sudden headache, nausea and vomiting. Computed tomography (CT) demonstrated a diffuse SAH, which was denser at the level of the right perimesencephalic cisterns, and a bilateral absence of the carotid canal in the bone window (Figure 1). Digital subtraction angiography (DSA) revealed a bilateral agenesis of the ICA (Figure 2), the fact that the carotid territory was supplied by elongated posterior communicating arteries (Figure 3), and a saccular aneurysm of the third middle of the basilar artery, which was completely occluded (Figure 4), but did not cause any neurological deficit. The aneurysm was treated via an endovascular route with detachable coils. For the safety of the procedure, balloon remodeling was placed, but inflation was not necessary, maintaining continuous distal flow.

Discussion

On the 24th day of ED, the third aortic arch begins to give rise to the common and proximal ICA [3]. Shortly thereafter, the cephalic portion of the dorsal aorta joins to the distal extremity of the third aortic arch and forms the distal part of the ICA. By about the sixth week, the common, internal and external carotid arteries are almost completed formed [4].

The failure of the third distal aortic arch to develop, or its incomplete development, results in agenesis or hypoplasia of the ICA, respec-

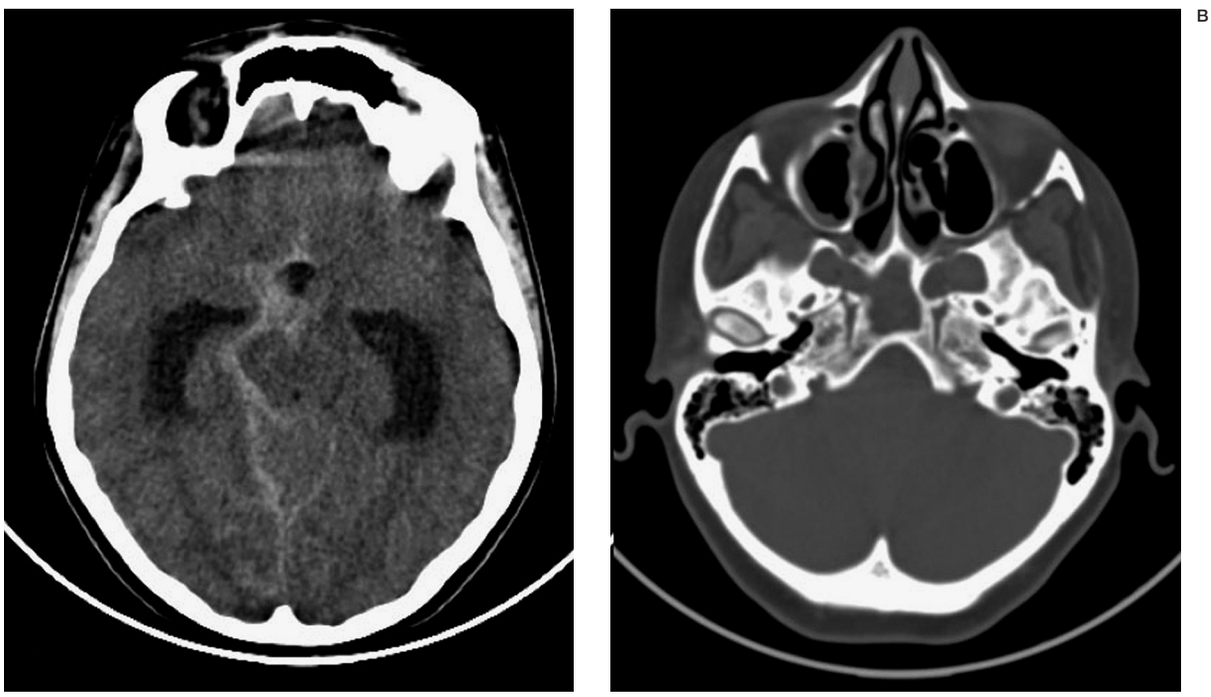

Figure 1 Unenhanced Axial CT shows a diffuse SAH, which is denser in the right perimesencephalic cisterns. Axial CT of the skull base bone window demonstrates the absence of the bilateral carotid canal.

tively, and can occur in unilateral or bilateral forms [4].

In this context, the absence of the ICA includes cases of agenesis, aplasia and hypoplasia. In such cases, collateral circulation is achieved principally through the circle of Willis (CW). Less commonly, the collateral flow is provided by persistent embryonic vessels, or by branches originating in the external carotid artery (ECA) [5].

The CW is formed when the embryo is between 7 and 24 mm. When the ICA is absent, the pattern of collateral circulation to the distal ICA and the intracranial vasculature depends on the stage of development at the time of the injury. The primitive routes of collateral circulation (i.e., the intracavernous anastomoses) will be overwhelmed if the injury occurs before the formation of the CW. In the same manner, collateral flow through the CW results if the injury occurs after the 24 mm stage of embryonic development [5,6].

Lie described six collateral circulation pathways associated with the absence of the ICA [7]. In type A cases, the unilateral absence of the ICA is associated with collateral circulation to the ipsilateral anterior cerebral artery (ACA) through the anterior communicating artery (ACOA), and to the middle cerebral artery (MCA) through a hypertrophied posterior communicating artery (PCOA). In type B cases, the ipsilateral ACA and MCA are supplied through the ACOA. Type C cases involve bilateral agenesis of the ICA, and the supply of blood to the anterior circulation, which is normally provided by hypertrophy of the PCOA, occurs via carotid-vertebrobasilar anastomoses. Type D cases involve unilateral agenesis of the cervical portion of the ICA and intercavernous communication of the ipsilateral carotid siphon to the contralateral cavernous ICA. In type E cases, small ACA's are supplied by bilateral hypoplastic ICAs, while the MCAs are supplied by enlarged PCOAs. In type F cases, collateral flow to the distal ICA occurs through transcranial anastomoses via branches of the internal maxillary artery [6-10].

A simplified classification scheme by Lie consists of three principle types of collateral circulation: collateral flow through the CW (most frequent), collateral flow via persistent fetal circulation, and reconstitution of the ICA through the skull base by external carotid artery [2,11].

An evaluation of the skull base and the presence or absence of the carotid canal is necessary to distinguish between agenesis and hypoplasia from an acquired condition. The presence of the ICA is a prerequisite for the development of the carotid canal. Therefore, the absence of the canal seen in a CT of the skull base confirms agenesis [12].

Two plausible causes for the increased prevalence of aneurysms in these patients have been put forward. They are the increased flow in the collateral vessels and the change in the vessels' dynamics [1,12]. Two mechanisms have been postulated to explain the strong association between intracranial aneurysm and agenesis/hypoplasia of the internal carotid artery: 1) both conditions could occur independently during embryonic life as the result of an error during development [13], or 2) the aneurysm may develop secondary to the hemodynamic disorder [1,14,15]. However, the increased prevalence of aneurysms among individuals with aplasia/hypoplasia of the ICA after the third decade of life suggests an acquired etiology. Zink et Al described that the large caliber of the collateral intracranial vessels observed in aplasia/hypoplasia of the ICA predisposes them to abnormal flow, and in turn to the development of aneurysms [3].

Our patient presented a saccular aneurysm that burst in the third middle of the basilar artery and agenesis of bilateral internal carotid artery, with collateral flow to the carotid territory by hypertrophied posterior communicating arteries (Type C Lie) (Figure 3) and small anastomosis between the ECA and right ophthalmic artery (Figure 2). The location of the aneurysm (the basilar trunk) is unusual and the association with primitive trigeminal artery was not observed in the DSA. This association is unlikely because we did not observe hypertrophy of the basilar artery, which is common in the presence of the primitive trigeminal artery [16].

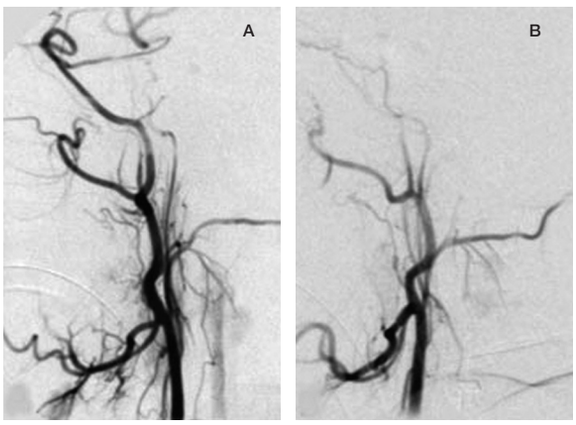

Figure 2 Lateral cervical projections of the right and left DSA of the ICA that demonstrate the absence of the internal carotid arteries small anastomosis between the ECA and right ophthalmic artery.

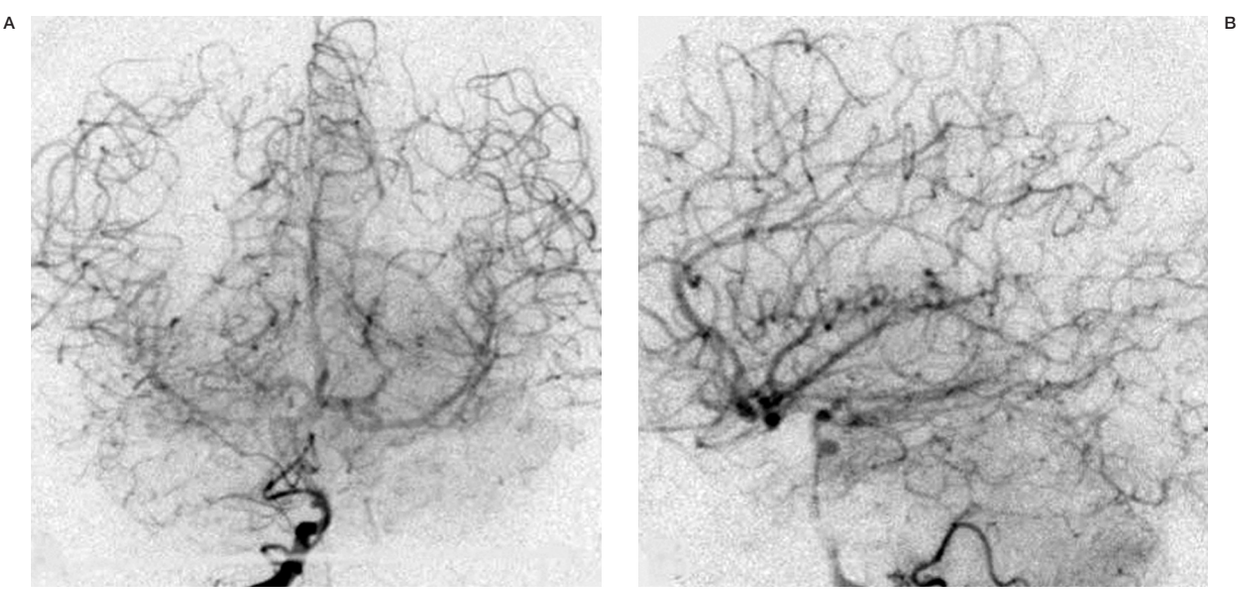

Figure 3 Anterior-posterior and lateral DSAs of the right vertebral artery that demonstrate opacification of the bilateral carotid artery through the posterior communicating arteries.

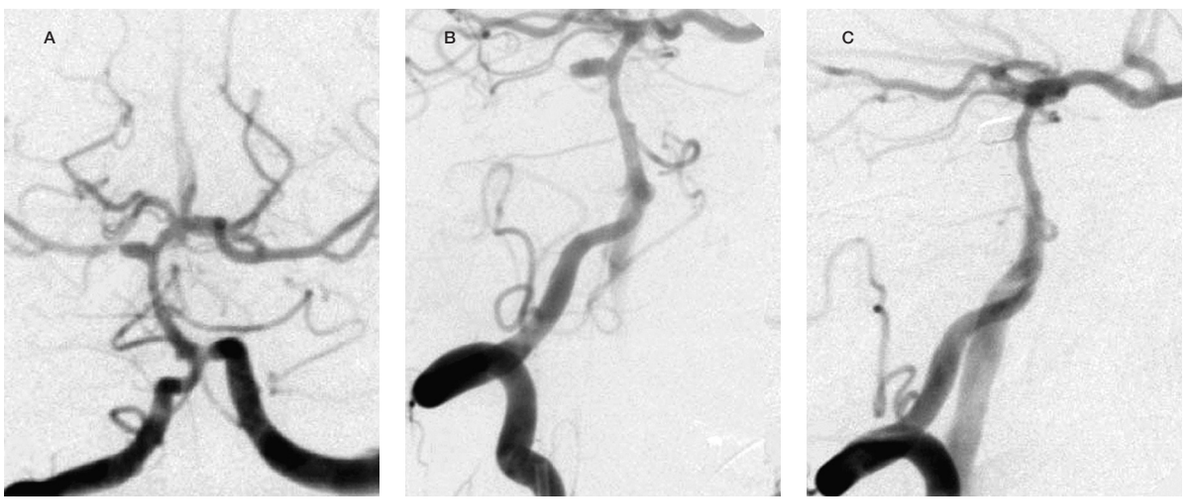

Figure 4 Anterior-posterior and oblique DSAs of the right vertebral artery pre-embolization that show the saccular aneurysm in the third middle of the basilar, and an oblique DSA of the right vertebral artery post-embolization that shows the coil packing with complete exclusion of the aneurysm.

We believe that despite the aneurysm not being located in a flow dynamic stress position, it is due to the hemodynamic disorder (increased flow in the basilar artery). An endovascular route was chosen as the means to effect treatment because it was the method with the lowest risk of morbidity or mortality due to the location of the aneurysm and the pattern of cerebral blood flow. The aneurysm was treated with detachable coils without complications. This treatment provided optimal angiographic and clinical results.

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